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CASE REPORT

Extranasopharyngeal angiofibroma: Report of two cases

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Abstract Nasopharyngeal angiofibromas are rare vascular tumours seen in ENT practice. Treatment of these tumours is a challenge for the ENT surgeon due to their high vascularity. They are mostly seen in male adolescents and are located in the nasopharynx with their origin being the sphenopalatine foramen. Extra-nasopharyngeal angiofibromas are very uncommon. We present here 2 such patients along with review of literature of extranasopharyngeal angiofibroma. One patient is a 28 years old where the tumour originated from the frontal recess, posterior to attachment of axilla of the middle turbinate. The second patient is a 12 year old boy where the mass was seen arising from the nasal septum. The unusual age of the patients and site of origin of these tumours make these unique cases worth reporting.

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1. Introduction

Juvenile nasopharyngeal angiofibromas are invasive, fibro-vascular tumours that occur almost exclusively in male adolescents. They originate from the region of the sphenopalatine foramen and enlarge to fill the postnasal space. Because of the close proximity to the nasopharynx and the ease with

which they invade this site, they are also commonly referred to as nasopharyngeal angiofibromas.¹ Nasopharyngeal angiofibromas may also extend to involve the sphenoid sinus superiorly, the nasal cavity anteriorly and the pterygopalatine fossa laterally.

Angiofibromas that do not originate from the area around the sphenopalatine region are rare and are referred to as extra-nasopharyngeal.^{2,3} In 1980, De Vincentiis and Pinelli reviewed a series of 704 cases of angiofibromas and found that 13 cases manifested outside the nasopharynx, thus suggesting that extra nasopharyngeal localization of this tumour is a possible, although rare occurrence.⁴ Since then, numerous studies have reported cases of angiofibroma localized in unusual sites. A recent review of the literature reported a total of 65 patients with atypical localizations of nasopharyngeal angiofibromas, the maxillary sinus being the most commonly involved site. Besides the different location, typical clinical characteristics of extra nasopharyngeal angiofibromas, such as, symptoms,

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age, sex, do not conform to a great extent with that of nasopharyngeal angiofibromas.⁵ Due to these different features, extra nasopharyngeal angiofibromas can present a diagnostic challenge and a meticulous evaluation with a high index of suspicion is essential in establishing the correct diagnosis and treatment.

2. Case reports

2.1. Case 1

A 28 year old male presented to the ENT OPD with left sided nasal obstruction and nasal bleeding for the past 9 months. Nasal obstruction was gradual in onset, progressive and was not relieved by medications. This was also associated with left nasal bleed, intermittent, moderate in amount and bright red in colour. Bleeding was controlled by digital pressure and the patient did not require nasal packing. There was history of hyposmia. Anterior rhinoscopy revealed a solitary reddish polypoidal mass seen in the left nasal cavity filling up completely and just short of the ala and pushing the septum to the right. On attempting decongestion, there was no decrease in size of the swelling. Nasal endoscopy was done to ascertain the findings. The mass was found to be attached just posterior to the attachment of axilla of the middle turbinate. The origin of the mass could not be confirmed as probing was not done. Contrast enhanced computed tomography (CECT) imaging of the nose and paranasal sinuses was suggestive of an enhancing nonexpansile mass in the left nasal cavity pushing the septum to the right side (Fig. 1A and B). The mass was blocking the left maxillary ostium with a collection of secretions in the sinus. The sphenopalatine foramen area was normal with no widening. With these findings a provisional diagnosis of a vascular tumour of the left nasal cavity was made and the patient was planned for endoscopic excision. Intraoperative on probing tumour bled profusely and required nasal packing. The mass was free from septum, lateral wall and floor and was seen to be arising from the frontal recess posterior to axilla of middle turbinate and it was removed in toto. There was a loss of about 50 cc of blood. Nasal packing was done and the packs were removed after 2 days. Biopsy report of the specimen

was suggestive of angiofibroma (Fig. 2). A postoperative CT scan shows no residual mass (Fig. 3A and B). Patient has been on regular endoscopic follow up for the last 6 months with no evidence of recurrence (Fig. 4).

2.2. Case 2

Twelve year old male patient presented with history of left nasal obstruction from 6 weeks. The nasal obstruction was insidious in onset, gradually progressive associated with bleeding from left side of the nasal cavity from 4 weeks. The bleeding was sudden in onset, episodic, profuse in amount and stopped by its own. There was no history of systemic bleeding, blood transfusion and nasal packing. On examination a single, smooth pinkish mass filling the left nasal cavity completely starting from the vestibule was found (Fig. 5A). CECT scan of the nose and paranasal sinuses showed contrast enhancing homogenous soft tissue density in the left nasal cavity starting anteriorly from the vestibule and posteriorly till 3 cm in the nasal cavity (Fig. 5B and C). In view of these findings differential diagnosis of hemangioma, angiomatous polyp and pyogenic granuloma was made. Endoscopic excision of mass was done under general anaesthesia. The mass was arising from septum just posterior to mucocutaneous junction and was removed in toto. The biopsy showed thin walled and angulated blood vessels and the blood vessels were lined by a single layer of endothelial cells and showed an incomplete muscular wall which was suggestive of nasopharyngeal angiofibroma (Fig. 5D). The patient is under follow-up and disease free.

3. Discussion

Nasopharyngeal angiofibroma is a well-known tumour of male adolescents. The tumour virtually always arises from the nasopharynx and only later may extend into the nasal cavity. More recently, the term extra nasopharyngeal angiofibroma has been applied to vascular, fibrous nodules occurring outside the nasopharynx.^{1,2} Angiofibromas that do not arise from the vicinity of the sphenopalatine foramen or pterygoid plates are referred to as extra-nasopharyngeal. Compared to naso-

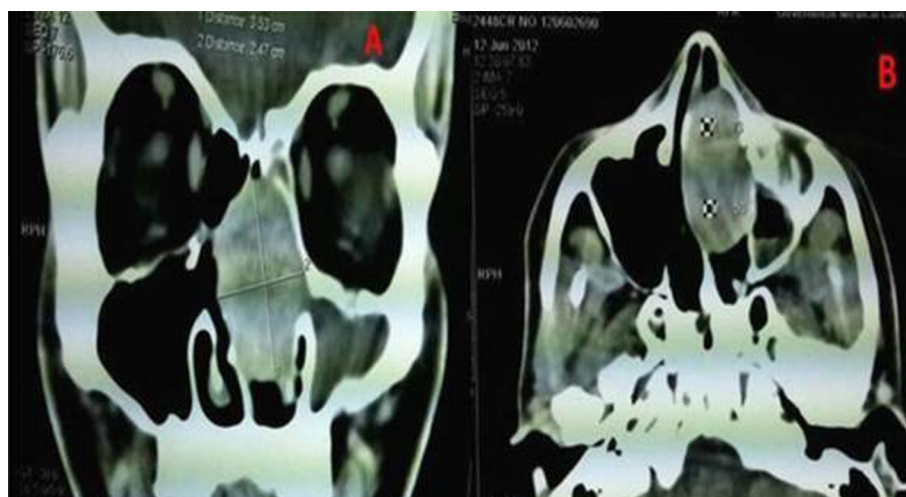


Figure 1 CECT nose & PNS showing a contrast enhancing mass in the nasal cavity, axial cut (A) & coronal cut (B).

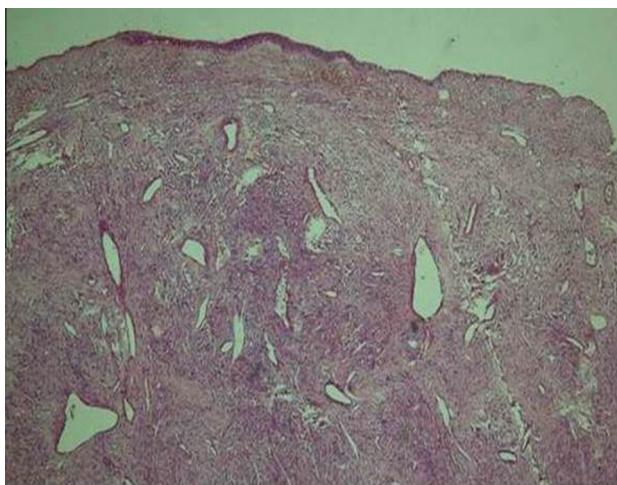


Figure 2 H&E, 100x. Stratified squamous mucosa with subepithelium showing admixture of thin walled blood vessels and fibrous stroma.

pharyngeal angiofibromas, patients affected are older, females can also be involved, symptoms develop more quickly, and hyper vascularity is less common.⁶⁻⁸ Juvenile nasopharyngeal angiofibromas are benign, unencapsulated, fibro-vascular lesions that account for less than 0.5% of all head and neck tumours.

These highly vascular tumours invade the nasopharynx, nose and skull base and commonly present with epistaxis and nasal obstruction. The most common primary extra-nasopharyngeal site for these tumours is the maxillary sinus.⁹ Other primary extra-nasopharyngeal sites reported are ethmoid and sphenoid sinuses, nasal septum, frontal recess, middle and inferior turbinates, tonsil, parapharyngeal space, ear, trachea, larynx, middle cranial fossa, infratemporal fossa, tonsil, retro molar region and conjunctiva.¹ The mean age of presentation of the tumour in extra nasopharyngeal sites is 22 years and the male to female ratio is roughly 3:1. In contrast nasopharyngeal angiofibroma presents almost exclusively in adolescent males with a mean age range between 14 and 17 years; female presentation is very rare.¹⁰ It is unclear why this discrepancy in the age of presentation and sex predilection exists between the two different types of angiofibromas. Contrast enhanced CT

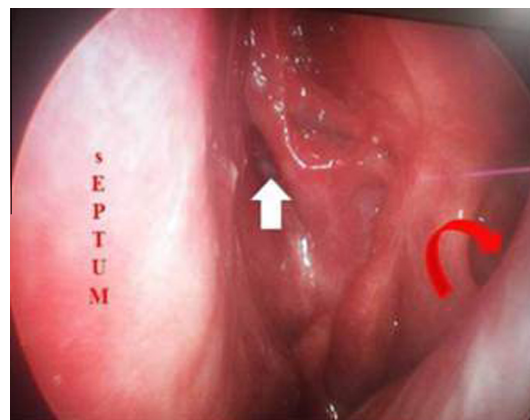


Figure 4 Post-operative endoscopic photograph showing a healthy cavity. White arrow pointing sphenoid ostium and curved arrow showing widened maxillary ostium.

scan (CECT) and magnetic resonance imaging (MRI) are used to determine the tumour site and its extension, with special attention being focused on skull base involvement, intracranial spread and relationship to important vascular and neurologic structures.^{2,9} While bone erosion can be more easily revealed by CT scan, MRI is adequate in demonstrating cortical erosion and cancellous replacement by tumour. Enlargement of the sphenopalatine foramen with erosion of the pterygoid plates are regarded as pathognomic radiological features and are best seen on CT. T1-weighted MRI will show a typical “salt and pepper” appearance caused by the increased vascularity of the tumour. Alvi et al. considered CT scan to be sufficient for the diagnosis of extra nasopharyngeal angiofibroma, as it clearly delineates and identifies the tumour.² However, signs of suspected hyper vascularity, upon CT scan or MRI, indicate the need for arteriography prior to surgical procedures in order to arrange the necessary precautions, such as embolization, and reduce the risk of brisk bleeding during tumour removal. Surgical excision of the mass is the treatment of choice, and recurrence is rare. Several authors have advocated different approaches for the surgery. Huang et al.⁹ and Handa et al.⁷ described lateral rhinotomy approach for tumours limited to

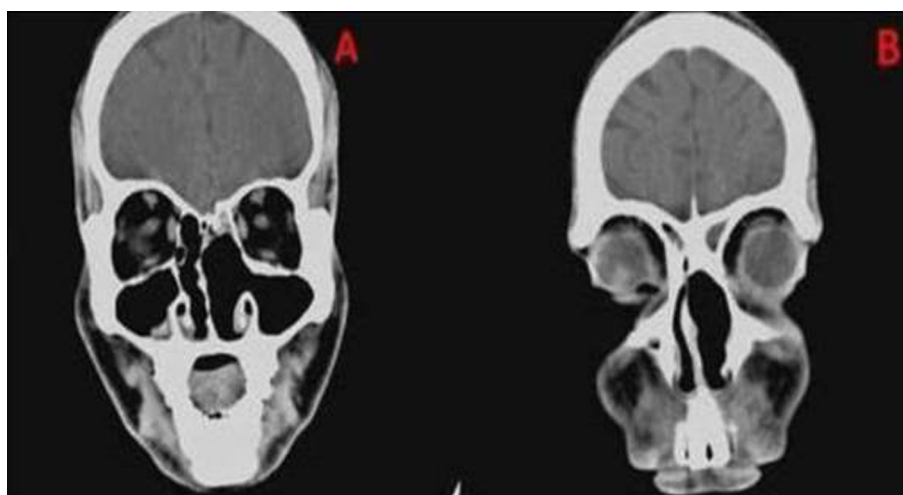


Figure 3 Post-operative CT scan axial cut (A), & coronal cut (B) showing complete removal of mass.

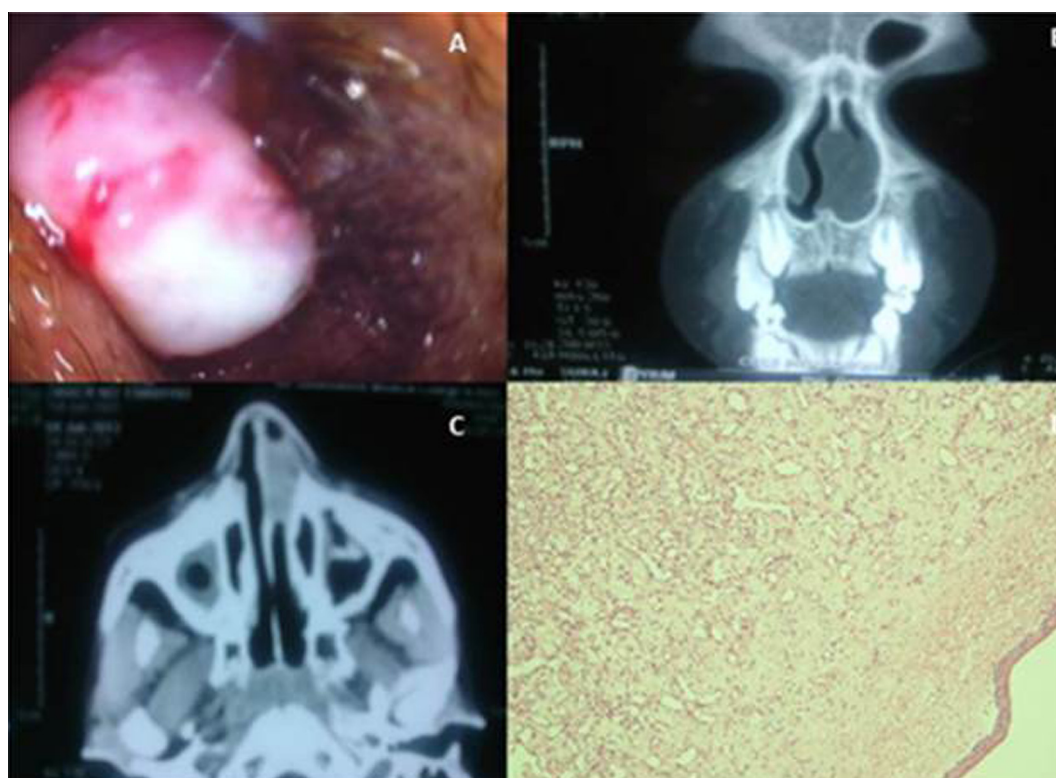


Figure 5 Pinkish mass filling left anterior nasal cavity completely (A), CT scan axial & coronal cuts showing mass in the anterior nasal cavity (B and C), histopathological picture showing thin walled vessels lined by single layer of endothelial cells with incomplete muscular wall (D).

the lateral wall of the nose. Manjalay et al.¹¹ used sub labial approach with carbon dioxide laser to remove the tumour. Hazarika et al.⁸ have described the use of laser and endoscope for the same. Advances in endoscopic Sino nasal surgery and the ability to embolize these tumours preoperatively have made many of the resections amenable to endoscopic technique. The decision to perform JNA resection endoscopically should be based on the experience and skill of the surgeon as well as the extent of the tumour (i.e. the lateral extent of the tumour must be accessible endoscopically). The surgeon must also be willing and capable to convert to an open approach if necessary. The morbidity of open approaches must be compared to the morbidity of incomplete tumour resection by performing the procedure via an endonasal approach. Patient selection for endoscopic resection is of paramount importance for a successful outcome. It has been suggested that tumours involving the ethmoid, maxillary, or sphenoid sinus, the sphenopalatine foramen, nasopharynx, or pterygomaxillary fossa and having limited extension into the infratemporal fossa are amenable to endoscopic resection. Complete excision is therefore undertaken for both therapeutic and diagnostic purposes. Extranasopharyngeal angiofibroma must be taken into consideration in the differential diagnosis of nasal vascular tumours and the nasal septum should be regarded as a potential, though exceptional, site of origin for these neoplasms.

Conflict of interest

None declared.

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